EXTENDED REPORT

Pellucid marginal corneal degeneration: evaluation of the corneal surface and contact lens fitting

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Aim: To quantify corneal irregularities, to describe the fitting with contact lenses, and to answer the question whether or not contact lenses with a special back surface design could improve visual acuity in patients with pellucid marginal corneal degeneration (PMCD).
Methods: 13 eyes were fitted with contact lenses with a special back surface. Videokeratographic data were assessed. The patients were followed up for an average period of 22.2 months. Lens tolerance and corrected visual acuity were evaluated.
Results: The mean eccentricity did not exceed 0.7 in all patients. Either the superior or the inferior eccentricity, or both, were negative in all patients. Using Fourier analysis all PMCD subjects showed an increased irregular astigmatism of the anterior cornea. Using Zernike coefficients seven eyes (53.8%) had a higher order aberration root mean square error (HOA RMS error) out of the normal range. The visual acuity with contact lenses improved in all eyes with an average increase of 2.7 lines (maximum eight lines). No serious complications were observed.
Conclusions: Quantitative evaluation of videokeratographic data may help to diagnose PMCD and to distinguish PMCD from other ectatic corneal diseases. Contact lenses with a special back surface design can improve visual acuity and lens tolerance.

Pellucid marginal corneal degeneration (PMCD) is a bilateral progressive ectatic corneal disorder characterised by a non-inflammatory inferior peripheral band of thinning extending from the 4 o'clock position to the 8 o'clock position. A 1–2 mm margin of normal cornea lies between the thinning and the limbus.[1 2]

The epithelium of the thinned out area is intact with an ectatic cornea superior to the thinning. The area between limbus and the thinning is clear, without any scarring, lipid deposition or vascularisation. Eyes may be affected asymmetrically and rarely the degenerative area may be located superiorly.[1 2]

Patients present in their third to fifth decade of life with a decrease of visual acuity because of high and irregular astigmatism. Videokeratoscopic colour coded maps show a marked flattening of the cornea along a vertical axis and a steepening of the inferior cornea peripherally to the site of the lesion resulting in a clinically irregular and against the rule astigmatism.[3 4] Reported complications are hydrops[5] and spontaneous perforation.[6]

Surgical options to improve the irregular corneal conditions include intrastromal corneal rings, thermocauterisation, and a great variety of different types of keratoplasty.[7–10] None of the present methods of treatment reliably achieve an optimum visual result. Thus, surgical intervention should be delayed as long as possible.

The non-surgical approaches to the management of the high and irregular astigmatism in patients with PMCD include spectacles and different groups of contact lenses.[11–13]

Patients with high or irregular astigmatism cannot be corrected with spectacles. In these cases contact lens fitting is a good option.[12 13] Among the different kinds of contact lenses a rigid gas permeable lens may be the correction of choice, since this type of lens provides good visual acuity, corrects high degrees of regular and irregular astigmatism, has high oxygen permeability, and, in comparison with soft contact lenses, carries a lower risk of microbial keratitis (incidence 1/10 000) and corneal neovascularisation.[14 15] Because of the special conditions, leading to changes of the inferior corneal surface, in most cases only rigid gas permeable contact lenses with a special back surface design can lead to optimal fit and visual results.

The aim of this study was to evaluate the special corneal conditions in patients with PMCD using videokeratoscopic indices and to clarify which special back design may be used to improve contact lens tolerance and visual acuity in cases of PMCD.

PATIENTS AND METHODS

Anterior surface of the cornea

The anterior surface of the cornea has a complex shape.[16 17] To analyse this shape and measure corneal irregularities videokeratographic colour coded maps, different parameters, such as Fourier series harmonic analysis[18 19] and Zernike polynomials,[20 21] have been developed.

Fourier series harmonic analysis

Using Fourier series harmonic analysis we can decompose corneal topography data into a series of trigonometric functions and thereby quantify each component such as the spherical power, the regular astigmatism, asymmetry, and higher order irregular astigmatism.[22 23]

Dioptric powers on a mire ring i, $F_i(\sigma)$ were transformed into trigonometric components of the following form[22]:

$$F_i(\sigma) = a_0 + c_1 \cos(\sigma - \alpha_1) + c_2\cos2(\sigma - \alpha_2) + c_3\cos3(\sigma - \alpha_3) + \ldots + c_n\cos n\,(\sigma - \alpha_n)$$

The resulting components of all rings were regrouped and displayed in separate images, where zero order (a_0) is the spherical equivalent, first order ($2c_1$) is the asymmetry component (tilt or decentration), second order ($2c_2$) is the regular astigmatism component, and third and higher orders ($c_{3\ldots n}$) are the higher order irregularity component. The

Abbreviations: BCVA, best corrected visual acuity; CL, contact lens; HOA, higher order aberration; PMCD, pellucid marginal corneal degeneration; RMS, root mean square

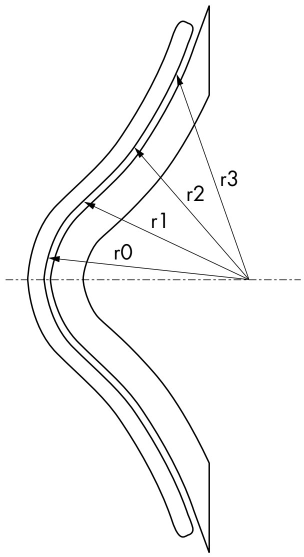

Figure 1 Keratoconus design. r^0 = central radius. r^1; r^2; r^3 = peripheral radii. The difference between r^0 and the peripheral radius r^2 is much greater than the central radius. r^3 is very flat.

normal range of the Fourier indices are defined as the mean plus or minus 2 SD in normal eyes as described by Tanabe *et al*, which are 40.81D–47.13D for spherical power, 0D–1.04D for regular astigmatism, 0.02D–0.68D for asymmetry, and 0.05D–0.17D for higher order irregularity.[23]

Zernike coefficients

From the Zernike coefficients, we calculated the root mean square (RMS) of higher order aberrations (HOA, square root of the sum of the squared coefficients of orders 3 up to 6) and the aberration coefficient (coefficients of order 1 to 8).[24 25]

The normal range of the HOA RMS error is defined as the mean plus or minus 2 SD in normal eyes as described by Wang *et al*, which is 0.100–0.670 μm.[26]

Eccentricity

The eccentricity factor describes the changing of the central corneal radius towards the periphery. The eccentricity in each corneal sector and a mean eccentricity were calculated. In healthy corneas the eccentricity lies between 0.5 and 0.7 in all corneal sectors.[27 28]

Contact lens fitting and anterior surface of the cornea

Depending on the central curvatures and the eccentricity of the corneas we selected the back surface design of our contact lenses. All contact lenses were made of rigid gas permeable (RGP) material. The following kinds of special back surface contact lenses were fitted.

Keratoconus design (fig 1)

This kind of back surface design can be fitted in early stages of PMCD because design and fitting is less problematic than bitoric design.

Backtoric design

The choice of backtoric or bitoric design can improve the fit of the contact lens and additionally correct remaining astigmatism especially in patients with a wide range of central corneal curvatures as can be found in PMCD patients with sophisticated corneal conditions.

Patients

Data evaluation

A retrospective chart review was conducted of all patients with PMCD who were fitted with specially designed contact lenses between November 2000 and January 2005. The data analysis of 13 eyes in nine patients, eight males and one female, included age, previous ophthalmic history, best corrected visual acuity with spectacles, postoperative best corrected visual acuity with contact lens, contact lens back surface design, and follow up time. Central corneal curvatures, eccentricities (ϵ), Fourier indices, and Zernike coefficients were performed by the Oculus computerised videokeratoscope (Oculus, Wetzlar, Germany; Software Version 1.64).

Diagnosis

The patients with PMCD were diagnosed after slit lamp examination, in which we found a typical thinning of the inferior peripherial cornea with a margin of normal cornea between the thinning and the limbus and corneal ectasia superior to the thinning. In all patients the topography showed "against the rule" astigmatism with an inferior steepening and a butterfly pattern (smiley or kissing birds) along the nasal and temporal hemimeridians (see fig 3A and B).

Patient profile

Table 1 shows the profile and the classification of the 13 eyes and the kinds of contact lenses fitted. In none of these patients could we fit a contact lens with regular back surface design because of the corneal conditions. The mean follow up time was 22.2 months. Depending on the corneal curvatures and on individual conditions we fitted 10 bitoric contact lenses and three contact lenses with a keratoconus design. The diameter of all contact lenses was 9.75 mm.

RESULTS

Table 2 summarises the keratometric data of all patients. All patients had an increased irregularity of the anterior cornea (fig 2).

Figure 3 demonstrates the deviation of the sectorial eccentricities in each patient. While both the nasal and the temporal eccentricities were greater than 0.63 in all patients, either the superior or the inferior eccentricity was negative in all patients. In two patients both superior and inferior eccentricities were negative. While the mean deviation between ϵ_{nas} and ϵ_{tem} was 0.12 (SD 0.07), the mean deviation between the nasal or the temporal eccentricity and the superior and the inferior eccentricity was between 1.05 and 1.12, the mean deviation between superior and inferior eccentricity was 1.06.

Using Fourier series harmonic analysis six eyes (46.2%) had spherical power out of the normal range, all eyes had central regular astigmatism out of the normal range (fig 4), and all eyes (100%) had irregular astigmatism (asymmetry and higher order irregularity) out of the normal range.

While decentration was out of the normal range in all eyes, higher order irregularity was out of the normal range in nine of 13 eyes (69.2%).

Using Zernike coefficients seven eyes (53.8%) had HOA RMS error out of the normal range. Five out of these seven eyes had a deviation of the central corneal curvatures >1 mm. The aberration coefficient,[25] which was calculated by using Zernike coefficients (first to eighth order), was out of the normal range in all eyes.

Table 3 demonstrates the data of contact lens fitting and the visual acuity of all patients.

The mean eccentricity was positive in 12 out of 13 cases (92.3%) and did not exceed 0.7 in all patients. In 10 out of 13 patients (76.9%) the mean eccentricity was smaller than 0.5.

The deviation of the central corneal curvatures ranged from 0.3 mm to 3.74 mm. In five cases the deviation was greater than 1.0 mm.

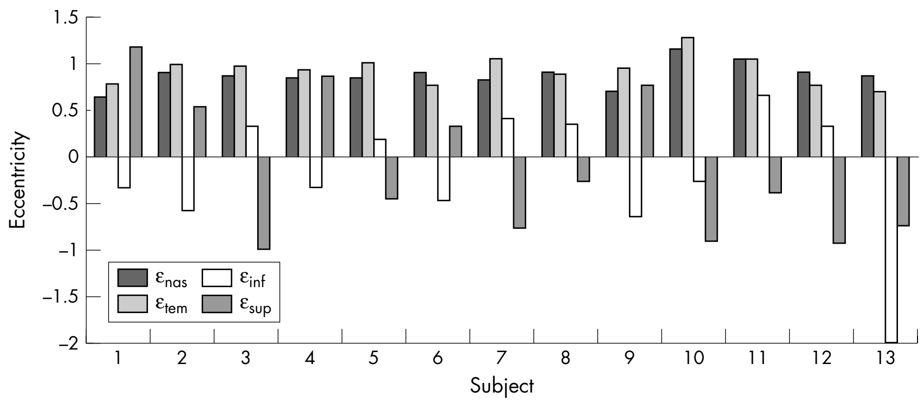

Figure 2 The diagram shows the sectorial eccentricities of all 13 subjects. The nasal as well as the temporal eccentricities are positive and have a mean deviation of 0.12 (0.07). Either the inferior or the superior, or both, eccentricities are negative with a mean deviation from the temporal eccentricity of 1.11 (0.63) and 1.12 (0.73), respectively. ϵ_{tem} = temporal eccentricity, ϵ_{nas} = nasal eccentricity, ϵ_{inf} = inferior eccentricity, ϵ_{sup} = superior eccentricity.

Either a bitoric design or a keratoconus design was necessary in all patients because of corneal conditions.

Only in three patients with deviation of corneal curvatures of less than 1 mm could a keratoconus design be fitted. The base curves of the contact lenses in case of keratoconus design were a mean value between both corneal radii and had to vary only minimally depending on individual conditions.

When we used a bitoric design, we flattened the steeper radius to reach an optimal contact lens fit. The remaining astigmatism was corrected by an additional front toric design of the contact lens.

In all 13 cases a good visual acuity was achieved with a mean increase of 2.7 lines and good contact lens tolerance (CL tolerance without any discomfort used 6 hours/day).

In all patients (100%) it was possible to improve the visual acuity in comparison with best corrected visual acuity with spectacles. The minimum improvement was one line, the maximum improvement was eight lines. We were able to achieve a satisfactory contact lens fit and we did not observe severe contact lens complications during the follow up period.

DISCUSSION

To the best of our knowledge this is the first report on the measurement of the irregular anterior corneal surface in patients with PMCD.

Up to now all cases with PMCD were diagnosed based on the presence of corneal thinning with ectasia above or below the area of thinning and the typical findings in colour coded maps showing against the rule astigmatism and inferior steepening with a butterfly (smiley or kissing bird) pattern of steepening.[1–4]

Table 1 Profile and classification of the patients

Total number	9
Male	8
Female	1
Binocular	13
Age	Mean 41.5 years (min 26 years; max 60 years)
Follow up time (mean)	22.5 months (max 50 months; min 13 months)
Contact lens material	
Rigid gas permeable	13
Soft	0
Contact lens back surface design	
Bitoric	10
Keratoconus design	3

Age and follow up time and time between keratoplasty and first contact lens fitting.

The obvious reason for carefull screening of all corneal parameters carried out by videokeratoscopy are patients for whom LASIK or other refractive surgery is planned. In two cases with normal central corneal thickness and stable refractions the LASIK procedure resulted in progressive keratoectasia because of non-detected PMCD.[29] Furthermore, because of the more severe complications and the differences in the choice of the contact lens back surface

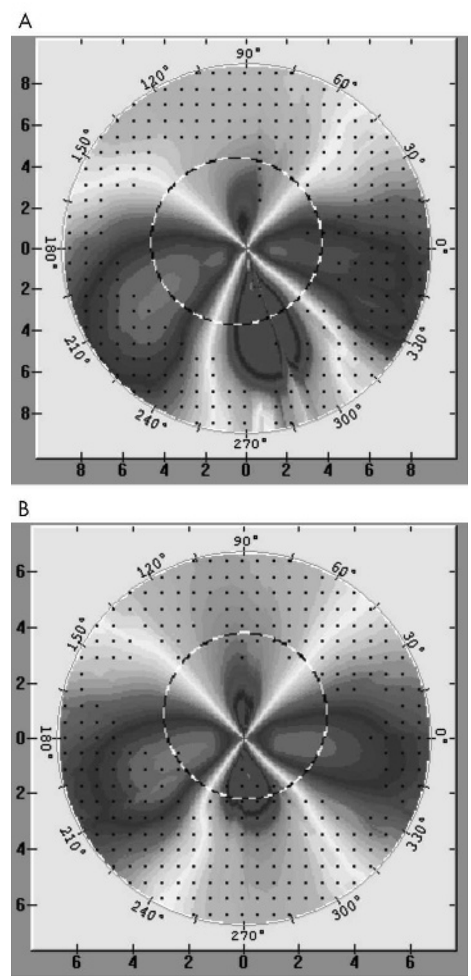

Figure 3 (A) Subject 11. (B) Subject 13. Colour coded maps of the videokeratopraphic data of both patients with inferior PMCD. Both show against the rule astigmatism with an inferior steepening and a butterfly pattern (smiley or kissing birds) along the nasal and temporal hemimeridians.

Table 2 Keratometric profiles of all subjects

			Fourier series harmonic analysis					Zernike coefficients	Aberration coefficient
			Spherical power	Regular astigmatism 3 mm	Regular astigmatism 6 mm	Decentration	Higher order irregularity	HOA RMS error	
			Normal range (D)[23]					Normal range (µm)[26]	Normal range[25]
No	**simK**	**Axis (°)**	**40.81–47.13**	**0–1.04**	**0–1.04**	**0.02–0.68**	**0.05–0.17**	**0.100–0.670**	**0–1**
Deviation of corneal radii <1 mm									
1	−2.2	115.7	*50.07*	*1.02*	0.69	*4.34*	*0.17*	0.324	*1.8*
2	−2	112.5	42.42	*2.04*	0.86	*2.86*	*0.26*	0.240	*1.5*
3	−2.6	66.9	46.73	*2.62*	0.80	*9.69*	0.12	0.216	*3.1*
4	−3.8	59.6	*48.48*	*1.47*	0.64	*3.47*	0.13	0.348	*1.7*
5	−3.4	70.4	43.24	*2.21*	*1.19*	*9.69*	0.14	0.763	*2.6*
6	−4.7	62.2	*50.00*	*1.86*	*2.27*	*2.15*	*0.53*	0.423	*2.0*
7	−3.8	116.6	43.64	*2.44*	*2.04*	*11.16*	0.15	0.823	*2.6*
8	−5.2	59.9	44.44	*3.41*	*5.52*	*2.86*	*0.42*	0.473	*1.8*
Deviation of corneal radii >1 mm									
9	−8	109.3	*51.77*	*1.64*	*4.47*	*10.50*	*0.38*	*1.029*	*3.2*
10	−11.5	64.3	43.64	*6.84*	*5.32*	*11.93*	*0.47*	*1.005*	*3.0*
11	−15.4	106.3	42.91	*13.26*	0.69	*2.09*	*0.91*	*0.964*	*6.6*
12	−12.8	97.5	*38.31*	*11.42*	0.86	*9.30*	*0.84*	*0.918*	*4.5*
13	−18.6	99.8	42.75	*10.91*	*0.80*	*2.15*	*2.11*	*1.314*	*3.5*
			Mean (SD)						
			45.26 (3.86)	*4.97 (4.62)*	*2.15 (1.94)*	*6.32 (4.01)*	*0.54 (0.58)*	0.672 (0.336)	*2.91 (1.4)*

All values out of the normal range are in italics.
All means of the Fourier series harmonic analysis without spherical power and of the aberration coefficient are out of the normal range.
The central regular astigmatism and the irregular astigmatism within the Fourier series harmonic analysis (asymmetry and/or higher order irregularity) are out of the normal range in all patients.
Regular astigmatism 3 mm, central regular astigmatism.
Regular astigmatism 6 mm, peripheral regular astigmatism.
HOA RMS error, root mean square error of the higher order aberrations (third to sixth order).

design in PMCD eyes in comparison with patients with other ectatic corneal diseases early detection of the PMCD might be beneficial.

To quantify these corneal changes we demonstrated the sectorial eccentricities, Fourier series harmonic analysis, the HOA RMS, and the aberration coefficient based on Zernike polynomials in patients with PMCD.

Contrary to patients with keratoconus, in whom mean eccentricity should be greater than 0.7 and positive in all sectors (fig 5),[27 28] we found a mean eccentricity that did not exceed 0.7 in all patients. The nasal as well as the temporal eccentricity was greater than 0.63 and either the inferior or the superior eccentricity was negative.

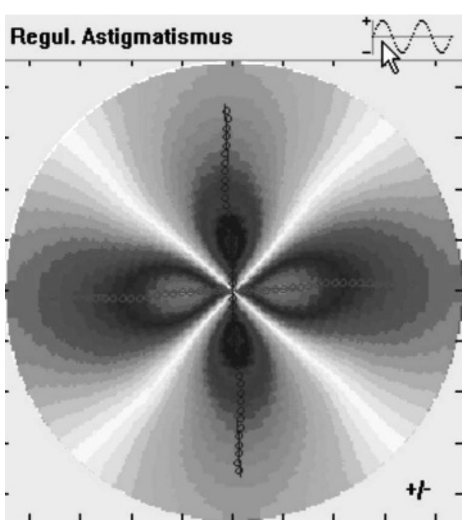

Figure 4 The colour coded videokeratographic map of the Fourier regular astigmatism of subject 12 shows the regular against the rule astigmatism with a typical trefoil pattern.

Using Fourier series harmonic analysis in all of the examined eyes the irregular astigmatism (asymmetry and/or the higher order irregularity) was out of the normal range.

Although in four eyes with corneal radii deviation <1 mm as well as asymmetry and higher order irregularity were out of the normal range, no increased HOA RMS was found.

Using colour coded maps of the Fourier regular astigmatism in PMCD subjects (fig 4) we found a regular against the rule astigmatism with a typical trefoil pattern contrary to subjects with keratoconus in whom a typical curl pattern with rotation of the peripheral astigmatism up to 90° has been seen (fig 2A and B).

In all five eyes with a deviation of corneal curvatures >1 mm all indices (asymmetry, higher order irregularity, HOA RMS) were out of the normal range.

In all eyes we found an increased aberration coefficient in comparison with healthy eyes.[25]

In our opinion sectorial eccentricites are the most valuable feature to analyse the corneas in case of PMCD followed by aberration coefficient and asymmetry.

However, aberration coefficient and asymmetry are not specific for PMCD and might also be increased in patients with keratoconus.[23 25] Colour coded maps of regular astigmatism might complete the videokeratoscopic analysis.

Publications on the fitting of contact lenses in patients with PMCD are rare. None of them describe the correlation between the anterior corneal surface and the choice of the back design of the contact lens.[12 13] Only one case report describes the use of a special design (reverse design) in a patient with PMCD.[11]

Table 3 Contact lens profile of all subjects. The selection of the lens design depends mainly on the central corneal curvatures

No	ϵ_{mean}	Corneal curvatures r_1	r_2	$r_2 - r_1$	CL design	Base curves		BCVA spectacles	BCVA CL
Deviation of corneal radii <1 mm									
1	0.57	6.69	6.99	0.30	Keratoconus	6.85		0.9	1.0
2	0.47	7.72	8.09	0.37	Bitoric	7.85	8.10	0.8	1.0
3	0.29	7.18	7.61	0.43	Bitoric	7.35	7.65	0.5	1.0
4	0.57	6.78	7.34	0.56	Keratoconus	7.15		0.9	1.0
5	0.41	7.54	8.16	0.62	Bitoric	7.75	8.20	0.4	1.0
6	0.39	6.53	7.19	0.66	Keratoconus	6.90		0.4	0.8
7	0.39	7.58	8.28	0.70	Bitoric	7.80	8.30	0.3	1.0
8	0.47	7.19	8.08	0.89	Bitoric	7.55	8.05	0.9	1.0
Deviation of corneal radii >1 mm									
9	0.45	6.40	7.53	1.13	Bitoric	6.95	7.85	0.6	0.8
10	0.32	6.98	9.16	2.18	Bitoric	7.70	9.20	0.4	1.0
11	0.6	6.52	9.29	2.77	Bitoric	6.80	9.30	0.5	1.0
12	0.27	7.60	10.69	3.09	Bitoric	8.20	10.70	0.7	1.0
13	−0.28	6.58	10.32	3.74	Bitoric	7.80	10.30	0.5	0.9

BCVA, best corrected visual acuity; CL, contact lens.
ϵ_{mean}, mean eccentricity of all sectorial eccentricites of each patient.
The BCVA CL improved in all subjects: does not depend on the deviation of the central corneal curvatures and the lens design.

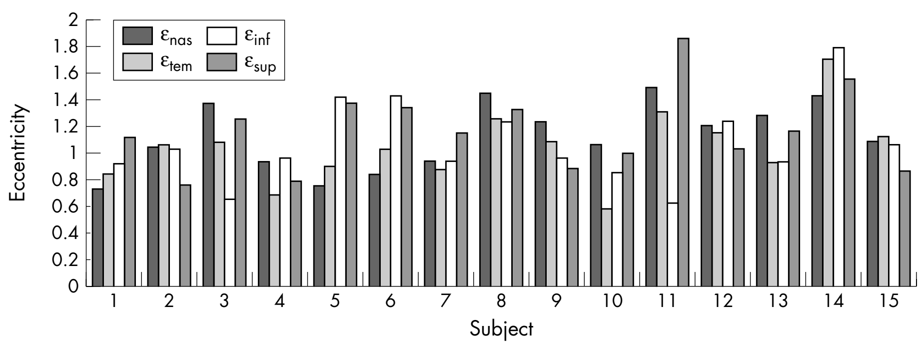

Figure 5 The diagram shows the sectorial eccentricities of 15 subjects with keratoconus. All eccentricities are positive and greater than 0.5. ϵ_{tem} = temporal eccentricity, ϵ_{nas} = nasal eccentricity, ϵ_{inf} = inferior eccentricity, ϵ_{sup} = superior eccentricity.

However, the anterior corneal surface in patients with PMCD can make it very difficult to achieve a sufficient correlation between anterior surface of the cornea and back surface of the contact lens and, as a result, improved visual acuity and good contact lens tolerance.

In most of the cases caused by the anterior cornea changes.

We were able to fit all of our patients with special contact lenses and to achieve improvement of BCVA with CL in comparison with the BCVA with spectacles and good contact lens tolerance. In none of the cases did we observe severe complications during the follow up period.

In our experience in cases of PMCD the choice of a bitoric design, depending on central corneal curvatures, leads to a reduction of the number of CL trials and to a shortening of the trial time and as a result to an increase in patient satisfaction.

The steeper central corneal radius should be flattened to improve the contact lens tolerance.

In cases of minor corneal radii deviation a keratoconus design might improve the situation.

Allthough mean eccentricity plays an essential part in contact lens fitting , because of the great variability of the sectorial eccentricites in patients with PMCD the role of eccentricity is minimised.

In our experience a reverse designed lens cannot improve contact lens fit in patients with PMCD. Reverse designed lenses should be fitted primarily in patients with negative eccentricities (for example, after penetrating keratoplasty). Here we demonstrate that in most PMCD patients the eccentricity was negative only in one sector. Therefore, an adhering of the reverse designed contact lens can result in a loss of tear film between the back surface of the lens and the anterior cornea.

Contact lenses are the most important alternative for correction of high or irregular astigmatism in patients with PMCD that involve minor risks and have good visual results. In our opinion, contact lenses with special back surface design can minimise problems in contact lens fitting and can improve the tolerance and the visual results. We recommend this procedure, especially for early and moderate PMCD, in cases of poor operative prognosis and for patients who refuse further surgical interventions. Furthermore, indices demonstrated here may supplement data for normal and pathological eyes[23 26] and, therefore, help detect suspected PMCD using computerised videokeratoscopy.

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